THE BRITISH JOURNAL
OF
CHILDREN'S
DISEASES.

REPRINT

PRINTED AND PUBLISHED BY ADLARD & SON,
BARTHOLOMEW CLOSE, LONDON, E.C.

ADDISON'S DISEASE IN A BOY, WITH CALCIFICATION OF THE ADRENALS, WITH REMARKS.

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Reprinted from 'The British Journal of Children's Diseases,' March, 1914, Vol. XI, pp. 105–111.

ADDISON'S DISEASE IN A BOY, WITH CALCIFICATION OF THE ADRENALS, WITH REMARKS.*

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A boy, aged $12\frac{3}{4}$ years, was bright, intelligent, and fair-skinned until about the end of October, 1913, when his skin was noticed to be brown; this was at first attributed to dirt and an attempt was made to wash it off. When this attempt failed it was thought to be due to his habit of drinking vinegar. Synchronously with his change in colour he lost all his energy, was inclined to sleep all day, very easily got tired, had a cough at night and nocturnal enuresis. There has not been any gastro-intestinal disturbance or fainting attacks. Except for chickenpox some years ago the patient has not had any illnesses. An uncle and a brother are said to have had tuberculosis. The other children and the parents are fair.

On admission the boy had a somewhat lifeless, sleepy, sunken-eyed appearance and a general bronzing of the skin, especially around the nipples, the umbilicus and pudenda. There is a large pigmented scar in the left groin, a smaller one in the middle line between the pubes and the umbilicus, and the back and abdomen show a few scattered spots of pigmentation. On the right side of the abdomen there are scars of a former herpes zoster, but these are not pigmented. The skin is somewhat dry and scurfy, and it is perhaps due to desquamation that a scratch on the skin remains white for a long time. There is no buccal pigmentation. There are no signs of pulmonary or spinal tuberculosis, and von Pirquet's reaction was negative on two occasions. The systolic blood-pressure in the arms varies between 64 and 84 mm. Hg. There is no valvular disease of the heart. There is no dulness behind the manubrium sterni. The spleen and liver are not palpable.

Skiagraphy: Dr. Stanley Melville reports—"There are very

* This case was shown on January the 9th, 1914, at the Clinical Section of the Royal Society of Medicine.

The treatment consisted in rest in bed and adrenalin chloride solution (1 in 1000), ten minims in water three or four times daily by the mouth. Later sandwiches containing raw adrenal glands were given. After this the pigmentation appeared to increase.

Remarks by Dr. H. D. Rolleston.

Addison's disease is rare in children. In 1897 Dezirot (4) collected 48 cases in children under sixteen years of age, and in 1910 Chemin (3) collected 56 cases under that age, but amongst these he includes the case of a girl, aged 3 years, which was published independently by Pitman (12), J. Ogle (11) and W. H. Dickinson (5), and was probably the earliest example of precocious sexual development associated with a cortical hypernephroma. Out of 55 of Chemin's cases 6 only occurred in the first decade, and J. Thomson (15) states that Addison's disease in children occurs almost exclusively over the age of twelve years. A most remarkable exception to this is the family with five cases of Addison's disease at the same time—in the mother, and four children aged 7, 4,* 3½,* 2½ years—recorded in 1900 from Edinburgh by Fleming and Miller (6). These patients when reported were all living, and the authors were obviously conscious of their exceptional nature, for they remarked that it may be questioned if these five individuals are all the subjects of true Addison's disease. It is noteworthy (*a*) that in "an account of the epidemic outbreak of arsenical poisoning in the north of England and in the Midland counties in 1900," E. S. Reynolds (13) mentions that some of the pigmented cases were diagnosed by his assistants as Addison's disease, though in describing the extent of the epidemic he does not mention Edinburgh or any town so far north; and (*b*) that Fleming and Miller's cases were published before the existence of this epidemic was recognised. Although there is no justification for considering Fleming and Miller's cases to be examples of arsenical pigmentation, the exceptional nature of the cases and the coincidence of the epidemic in another part of the country are certainly suggestive.

* These ages may remind the reader of the lines in James and Horatio Smith's 'Rejected Addresses' and their own footnote:

"My brother Jack was nine in May†
And I was eight on New Year's Day."

"† Jack and Nancy, as was afterwards remarked to the authors, are here made to come into the world at periods not sufficiently remote. The writers were then bachelors."

In Chemin's 55 cases, 32 were males and 23 girls. He concludes that the features of the disease in children, which differ from those seen in adults, are darkening of the hair, diarrhœa (present in the four children reported by Fleming and Miller), incontinence of urine, and the more rapid course of the malady. Darkening of the hair, however, may occur in adults. It is now generally recognised that Addison's disease may be complicated by the *status lymphaticus*; out of 15 cases of Addison's disease Hedinger (8) found that seven showed *status thymico-lymphaticus*, and five pronounced lymphatic hyperplasia. In Langmead's (10) case of Addison's disease in a boy, aged 10 years, the spleen, 5 oz., came down almost to the umbilicus. A high lymphocyte count, such as was seen in the two first blood examinations of our case, is regarded as evidence of lymphatism and therefore as a bad prognostic. The eosinophile count in our case was high—7, 6·5, 4, and 6 per cent.; Gulland and Goodall (7) refer to a case with 10 per cent. For the increased number of red blood-corpuscles occasionally seen in Addison's disease various explanations have been offered. It has generally been assumed that it is due to concentration caused by vomiting and diarrhœa, neither of which occurred in our case. A specific action of the tuberculous toxin has been invoked, and in this connection it may be mentioned that erythræmia has been recorded in about 6 out of 50 collected cases of primary massive tuberculosis of the spleen (Winternitz (18)). G. R. Ward (16) regards erythræmia in Addison's disease as a compensatory process, and argues that adrenal insufficiency gives rise to circulatory stasis, which in its turn causes concentration of the blood and actual increased formation of red blood-cells. The small size of the heart, shown by the *x* rays in our case, was insisted on by C. R. Box (2), and in Langmead's case of Addison's disease in a boy, aged 10 years, the heart weighed 3 oz. and resembled that of a child of two or three years old. The small size of the heart is no doubt closely correlated with the absence of adrenin and with the low arterial blood-pressure.

In an account of the pseudo-meningitic form of acute adrenal insufficiency, Sergent (14) described a cutaneous sign which he considered characteristic of low blood-pressure and adrenal insufficiency. On irritating the skin by a scratch a persistent "white line," due to reflex capillary spasm, results, which is the exact reverse of the well-known *tache cérébrale* in tuberculous meningitis. According to Le Clerc (9) this sign had long before been noticed in diseases other than Addison's disease by Gubler; and L. Bernard (1) brought forward evidence to show that it is valueless as an indication of low

blood-pressure and adrenal insufficiency. Our patient certainly presented it, but as already mentioned, it may well have been due to partial detachment of some of the dry epidermis.

There is a point of speculative interest in connection with the cutaneous scars: one in the left groin was much pigmented, a common event in Addison's disease; but on the right side of the abdomen and thorax there were scars, exactly like those of a former herpes zoster, which were pale and contrasted with the adjacent pigmented skin. In answer to the question why the latter were not pigmented, it may be suggested that the absence of pigment is connected with impaired innervation of the herpetic scars. If this is so, it is an argument in favour of the view that the bronzing of the skin in Addison's disease is due to nervous irritation.

The demonstration of calcification in the adrenals and in the bronchial lymphatic glands by skiagraphy is interesting, especially in so young a patient. It points to obsolete tuberculosis and thus agrees with the negative von Pirquet's reaction. Presumably cicatricial contraction in the calcifying adrenals, rather than active tuberculosis, is responsible for the recent appearance of symptoms of Addison's disease. As an exceptional example of calcification, though not subsequent to tuberculosis, attention may be drawn to Hale White and Bryant's (17) case of wide-spread arterial calcification and endarteritis associated with hydronephrosis in an infant of six months. In connection with Addison's disease and calcification reference may be made to the case of a man, aged 25 years, with cutaneous pigmentation, asthenia, which suggested Addison's disease during life, and wide-spread arterial calcification. At the necropsy the suprarenals were healthy, and the main visceral lesions were fibrosis of the left kidney and complete destruction of the right kidney by old disease. The myocardium showed calcification (B. Bramwell, 'Clinical Studies,' 1909, vii, p. 242).

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